

# Behavioural profile and maternal stress in Greek young children with Williams syndrome

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## Abstract

**Background** Williams syndrome (WS) is a genetic disorder causing intellectual disability. Children with WS often exhibit various kinds of maladaptive behaviours that affect their social functioning. In order to determine whether these behaviours are syndrome-specific, it would be necessary to compare children with WS with children with other syndromes as well as to provide data on the socio-emotional profile in WS from a variety of cultures. The present study investigated the behavioural profile and its relation to maternal stress in Greek young children with WS in comparison with young children with Down syndrome and typically developing (TD) children.

**Methods** Participants were 60 mothers, 20 in each syndrome group and 20 in the control group. The three groups were matched for mental age. The behavioural profile of the participants was investigated through the Child Behaviour Checklist (1.5–5 years) and maternal stress through the Parental Stress Index.

**Results** In accordance with studies in other cultures, it was found that young children with WS received significantly higher rates in emotional problems and anxiety/depression, compared with both children with Down syndrome and TD children. Moreover, mothers of children with WS reported significantly higher scores in the Total Stress index compared with mothers of TD children. However, in contrast with previous studies, only 25% of children with WS fell into the clinical range in the total Child Behavior Checklist score.

**Conclusion** The consistency of the socio-emotional characteristics of children with WS across cultures and developmental stages implies a strong influence of the genetic phenotype. However, Greek mothers avoided to characterize these behaviours as pathological. Implications of these findings for clinical practice are also discussed.

## Keywords

maladaptive behaviours,  
maternal stress,  
socio-emotional profile,  
Williams syndrome

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## Introduction

Williams syndrome (also Williams–Beuren syndrome) (WS) is a neuro-developmental disorder caused by the deletion of about 25 genes from the region q11.23 of chromosome 7 (Hillier *et al.*

2003). The prevalence of WS is estimated at about 1 in 7500 live births (Strømme *et al.* 2002). Physical characteristics of WS include specific facial anomalies, supravalvular aortic stenosis, and transient hypercalcaemia (Morris 2006). In the cognitive domain WS is characterized by mild-to-moderate intellectual

disability as well as profound deficits in visuo-spatial functioning, planning and problem solving (Arnold *et al.* 1985; Bellugi *et al.* 1990; Levitin *et al.* 2004). As regards the language domain, articulation, vocabulary, syntax and story telling are considered as areas of relative strength, while widespread problems are observed in abstract concepts of semantics, complex aspects of syntax as well as some pragmatic functions (e.g. Mervis 2006).

The socio-emotional phenotype of the condition has also been widely studied and individuals with WS are described as highly sociable, approaching, empathic and overfriendly even with strangers. Moreover, individuals with WS have been reported to suffer from fears and anxiety, attention problems and distractibility, while they often exhibit poor adaptability, low tolerance in frustration, negative mood and difficulties with peer relationships (Trauner *et al.* 1989; Udwin & Yule 1991; Gosch & Pankau 1994; Levine & Castro 1994; Wiegers *et al.* 1994; Van Lieshout *et al.* 1998; Mervis & Klein-Tasman 2000; Einfeld *et al.* 2001; Mervis *et al.* 2001; Semel & Rosner 2003). However, in order to identify whether this profile is syndrome-specific and highlight further the genotype-phenotype relationship it would be necessary to compare individuals with WS with CA and MA matched individuals with other syndromes as well as typically developing (TD) controls, to investigate the socio-emotional profile of WS individuals at different developmental stages, and to collect relevant data from diverse cultures.

As regards the first point, some studies suggest that there may not be socio-emotional characteristics specific to WS. Dykens and Rosner (1999) compared individuals with WS aged 14–50 years with equal numbers of age- and gender-matched individuals with Prader-Willi syndrome (P-WS) using the Reiss Profiles of Fundamental Goals and Motivation Sensitivities for Persons with Mental Retardation. Results showed that while there were significant differences across groups on 11 out of 15 items assessed, there were no characteristics specific to individuals with WS. In particular, the WS group was rated higher on the items ‘often initiates interactions with others’, ‘never goes unnoticed when in a group’, ‘has many fears’ and ‘feels terrible when others are hurt’. However, both the WS and the P-WS groups showed ‘strong desire to help others’ and ‘were very happy when others do well’. Contrarily, other studies demonstrate that individuals with WS appear very different from those of other syndrome groups or TD individuals. Tomc and his colleagues (Tomc *et al.* 1990) found that toddlers and school children with WS were rated by their parents significantly higher in approaching, intensity, distractibility and negative mood, but significantly lower in persistence and threshold of excitability compared with norms for TD children. Van Lie-

shout and colleagues (1998) examined children and adolescents with WS in relation to children and adolescents with P-WS, fragile-X syndrome (f-XS) as well as CA-matched controls. Their findings showed that the WS group received similar Extraversion and Agreeableness scores as normally developing group but it scored much higher on Agreeableness than both the P-WS and f-XS groups. Moreover, WS individuals scored lower on Conscientiousness, Emotional Stability, Openness, Motor Activity and Irritability than non-clinical individuals. Sarimski (1997) investigated the behavioural profile of children with WS in comparison with the behavioural profile of children with P-WS and f-XS using the Child Behaviour Checklist. All children aged 1–12 years. Some findings that appear to be specific to WS are that children with WS are more likely to be rated as worrying, talking too much and been overfriendly to strangers. On the other hand, attention problems are similar to those exhibited by children with f-XS, but significantly worse than those exhibited by children with P-WS. The authors concluded that this kind of problems is likely not specific to WS. Similarly, Gosch and Pankau (1994) used a list of 20 items of the Child Behavior Checklist (CBCL) and the Vineland Social Maturity Scale (VSMS) to compare children with WS and children with Down syndrome (DS). They found that children with WS were significantly less reserved towards strangers and more anxious, while they exhibited lower adaptability. In Doyle and colleagues (2004) study parents of children with WS, DS and normal controls provided data concerning specific aspects of their children’s social behaviour on the Salk Institute Sociability Questionnaire (SISQ). Children ranged in age from 1 to 12 years. Consistent with earlier findings, whole group analyses showed that the WS group was rated significantly higher on all aspects of sociability studied. Comparisons among the groups at different ages revealed that children with WS exceeded significantly children with DS on Global Sociability and Approach Strangers in every age group. Klein-Tasman and Mervis (2003) conducted a study comparing 8-, 9- and 10-year-olds with WS with a group of CA- and IQ-matched children with a variety of syndromes including DS. Two measures were used: the Children’s Behaviour Questionnaire (CBQ) and the Multidimensional Personality Questionnaire (MPQ). Results revealed that the distinctive characteristic of WS rests on the focus of these children on other people. The focus on others evidenced by individuals with WS includes also a component of anxiety related to social situations.

As regards the second point, general longitudinal studies have demonstrated that behaviour and emotional difficulties evidenced by individuals with WS persist over time (Davies *et al.* 1998; Udwin *et al.* 1998; Einfeld *et al.* 2001; Dykens 2003).

It has been observed that as individuals with WS grow older, they become less lively, determined, active, restless, tearful, quarrelsome, impertinent and overfriendly, but more depressive in comparison with younger individuals with WS. Additionally, adolescents and adults with WS are assessed as more withdrawn but better balanced than children with the condition (Gosch & Pankau 1997). However, most of the studies on the emotional and behavioural characteristics of WS include children and adolescents. On the other hand, there are relatively few studies regarding young children and even those do not focus on this developmental stage, but they include participants of a wide age range. The examination of behaviour problems in young children with WS would highlight further the role of the genotype in the expression of these problems, because at this developmental stage the role of socialization goals is still restricted. Moreover, a detailed description of the socio-emotional profile of young children with WS may contribute to the formation of more effective early intervention programs for the modification of maladaptive behaviours in this population.

In order to further investigate whether the emotional and behavioural characteristics of WS are genetically determined, it would be useful to study their expression in a variety of cultures. However, so far most of the relevant studies were carried out in western societies such as the USA, the Netherlands and Germany. Western societies are characterized as 'individualistic' and are dominated by a self-view that emphasizes uniqueness of the individual and leads to defining one's self in terms of one's own feelings and actions. Socialization goals comprise assertiveness, social competence with peers and courtesy (Triandis 1989). On the other hand, there are very few studies examining the emotional and behavioural characteristics of young children with WS coming from 'collectivist' societies. Collectivist societies are dominated by a self-view in which one defines one's self in terms of relationships with others. Socialization goals comprise obedience, gentleness and self-control. In this vein, Zitzer-Comfort and colleagues (2007) found that children with WS in Japan, like children with WS in the USA, exhibited overall more sociable behaviours and were more likely to approach strangers than their normal counterparts.

Thus, the investigation of the behavioural phenotype in Greek young children with WS may add to the relevant literature. Greek culture shows certain similarities but also considerable differences from other Western societies such as American, German and Dutch. Greek culture lies in between 'individualism' and 'collectivism' and primes an autonomous relational conception of the self who is simultaneously independent and interdependent. Socialization goals comprise

obedience and respect, but children are also praised for being unique and remarkable (Vassiliou 1966; Katakis 1976, 1978).

Despite the fact that the socio-emotional problems exhibited by individuals with WS may have genetic predispositions their course as well as any referral practices strongly depend on the maternal perceptions and reactions to these problems. Several studies have demonstrated that intellectual disability may be a source of stress in families of handicapped children (Turnbull *et al.* 1984; Friedrich *et al.* 1985; Byrne *et al.* 1988; Donovan 1988). Until the 90s it was believed that the main stressful factor in families of handicapped children is the degree of intellectual disability (i.e. mild, moderated, severe), regardless of aetiology (Hodapp & Dykens 1994). However, more recently it has been shown that family stress levels may also be affected by the degree and type of the handicapped child's socio-emotional problems, which are related to the aetiology of the condition (Gallagher *et al.* 1983; Margalit *et al.* 1989). Following this approach, some studies focused on the effects of different maladaptive behaviours, related to different genetic disorders, on maternal stress and classified participants by the aetiology of intellectual disability. In this vein, Fidler and colleagues (2000) compared stress levels in American families of young children with WS, DS and Smith–Magenis syndrome. Results showed that although parents in the WS group did not report more parent or family problems than the parents in the DS group and the Smith–Magenis syndrome group, they experienced more pessimism compared with the DS group. Sarimski (1997) compared stress levels in German families of young children with WS, P-WS and f-XS using the Parenting Stress Index (PSI). According to the findings, mothers in the WS group reported higher level of parent stress compared with mothers in the f-XS group, which resulted mainly from a higher level of depression. Moreover, child's distractibility and hyperactivity were more a problem for mothers of children with WS than for mothers of children with P-WS. Similarly, Van Lieshout and colleagues (1998) compared stress levels in Dutch families of children and adolescents with P-WS, f-XS and WS, and revealed that the parents of children with WS feel less capable of controlling the behaviours of their child but also less angry with their child than the parents of children with P-WS.

Taking into account the previous considerations this study has a twofold purpose: (i) to provide a comprehensive account of the behavioural profile of Greek young children with WS; and (ii) to examine the effect of this behavioural profile on maternal stress, in comparison with the effect of the behavioural profile exhibited by children with DS, which also causes intellectual disability, and matched TD children.

## Methods

### Participants

The participants of this study included 60 mothers of young children, 20 in each syndrome group and 20 in the control group. Young children were selected in order to obtain a homogenous sample and avoid any effects of the child's age on maternal responses. The three groups were matched for mental age using the Bayley Scales of Infant and Toddler Development (Bayley III) (Bayley, 2006). Information on children's mental and chronological age, gender and birth order is presented in Table 1. Thirteen children with WS (65%) and 12 children with DS (60%) received early intervention services by a speech therapist and an occupational therapist approximately three times a week. Only four children (20%) in each the WS and the DS group also received psychological support. As regards schooling, 12 children with WS (62%) attend private mainstream schools, while none of them attends special school. On the other hand, seven children with DS (35%) attend private mainstream schools and 7 (35%) attend public mainstream schools. Two children in this group (10%) attend public special schools. Eleven children in the control group (55%) do not attend any school, while six children (30%) attend mainstream private schools and 3 (15%) attend mainstream public schools.

Mothers' age and education in each group are also presented in Table 1. Mothers' age was on average 35.7 years (range 28–48 years). Twenty-seven percent of the mothers had finished high school, 39% had Technological Education and 22% were University educated. None of the mothers in either group was attending any counselling program for encountering child maladaptive behaviours.

**Table 1.** Demographic information for the Williams syndrome (WS), Down syndrome (DS) and the control group

	DS (n = 20)	WS (n = 20)	TD (n = 20)
Chronological age (in moths)*			
Mean (SD)	64.9 (15.1)	61.5 (17.7)	30.2 (8.9)
Mental age (in months)			
Mean (SD)	28.7 (6.2)	30.0 (10.0)	30.4 (9.8)
Gender			
Boys : girls	16:4	11:9	11:9
Birth order			
First born : later born	7:13	9:11	10:10
Mother's age			
Mean (SD)	36.4 (3.7)	36.0 (5.8)	34.7 (3.5)
Mother's education			
Mean (SD)	3.8 (0.9)	4.0 (1.1)	4.2 (1.1)

\* $P < 0.001$ .

TD, typically developing.

### Measures and procedure

The behavioural profile of the participants was investigated through the CBCL (1.5–5 years) and maternal stress through the PSI (Abidin 1995). Both questionnaires have been standardized in the Greek population.

The CBCL (1.5–5 years) is a parent-reported measure assessing common maladaptive behaviours in childhood. The Greek version of the CBCL consists of 100 items grouped in two 'wide-band' factors – internalizing and externalizing behaviours – and seven 'narrow-band' factors, namely emotional problems, anxiety/depression, somatic complaints, withdrawn, sleep problems, attention problems and aggression. Parents are asked to note whether each behaviour is 'not true' (0 points), 'sometimes true' (1 point) or 'very true' (2 points) for their child. The CBCL is commonly used to assess maladaptive behaviours in children with intellectual disability (Margalit *et al.* 1989; Hodapp *et al.* 1997, 2003; Sarimski 1997).

The PSI (Abidin 1995) was designed to measure parenting stress as a result of the parent–child interaction. The Greek version consists of 120 items grouped into a Child Domain and a Parent Domain. The Child Domain was constructed to examine child temperament factors that cause frustration to parents in trying to develop a relationship with their child, and includes the following subscales: adaptability, acceptability, demandingness, mood, distractibility/hyperactivity and reinforcement for parents. The Parent Domain reflects sources of stress regarding the parenting role that originate from the parent him/herself, and includes the following subscales: depression, attachment, restriction of role, sense of competence, social isolation, relationship with spouse and parental health. There is also a Total Parenting Stress Index, which gives an indication of the degree of the dysfunctional parenting behaviour, as well as an index of the amount of life stress experienced by the parent.

Mothers completed the questionnaires during a researcher's visit at home as part of a more extensive assessment of the child.

## Results

Group differences on the CBCL and the PSI were determined using a one-way ANOVA and Tukey's HSD *post hoc* tests. As regards the CBCL, results demonstrated significant differences on the subscales of emotional problems [ $F(2, 58) = 6.31, P = 0.005, \eta^2 = 0.26$ ], anxiety/depression [ $F(2, 58) = 6.53, P = 0.004, \eta^2 = 0.27$ ], withdrawn [ $F(2, 58) = 9.82, P < 0.001, \eta^2 = 0.36$ ], attention problems [ $F(2, 58) = 5.89, P = 0.006, \eta^2 = 0.25$ ], internalizing behaviours [ $F(2, 58) = 9.02, P = 0.001, \eta^2 = 0.34$ ] as

	Normative sample		DS		WS		TD	
	Mean	SD	Mean	SD	Mean	SD	Mean	SD
Emotional problems	2.87	1.77	1.64	1.50	3.75	2.30	1.52	1.50
Anxiety/depression	3.97	1.73	1.93	1.27	4.33	1.72	2.50	2.19
Somatic complaints	2.34	1.76	2.14	1.92	2.42	1.93	1.33	1.43
Withdrawn	1.87	1.42	2.43	1.45	3.08	2.15	0.42	0.67
Sleep problems	3.34	1.95	1.79	1.58	3.42	1.62	2.42	2.64
Attention problems	2.52	1.68	3.07	1.14	4.42	2.02	2.17	1.64
Aggression	10.43	5.10	7.79	4.19	11.17	5.72	8.42	5.40
Internalizing behaviours	10.71	4.92	8.14	4.05	13.58	5.45	5.67	4.54
Externalizing behaviours	12.94	6.27	10.86	4.88	15.58	7.10	10.58	6.83
Total score	37.34	15.47	31.43	13.51	46.58	17.53	25.83	15.38

\*Raw scores.

DS, Down syndrome; TD, typically developing; WS, Williams syndrome.

well as on the total CBCL score [ $F(2, 58) = 5.83, P = 0.007, \eta^2 = 0.25$ ]. As can be observed in Table 2, children with WS showed significantly higher rates compared with both children with DS and TD children in the subscales of emotional problems and anxiety/depression, in the 'wide-band' factor of internalizing behaviours as well as in the total CBCL score. Moreover, children with WS showed significantly higher rates compared with TD children in the subscales of withdrawn and attention problems. On the other hand, children with DS showed significantly higher rates compared with TD children only in the subscale of withdrawn.

In order to determine the percentage of children falling into the clinical or borderline range, the scores on the subscales and the 'wide-band' factors of the CBCL as well as the total CBCL scores were compared with norms established through the standardization of the CBCL in the Greek population. In the WS group 8 (40%) children scored in the clinical range or borderline range on the attention problems subscale, 5 (25%) on the withdrawn subscale, 3 (15%) on the aggression subscale and 3 (15%) on the somatic problems subscale. Moreover, 3 (15%) children in the WS group scored in the clinical or borderline range on the 'wide-band' factor externalizing behaviours, 2 (10%) on the 'wide-band' factor internalizing behaviours and 5 (25%) on the total CBCL score. On the other hand, only 3 (15%) children with DS scored in the clinical or borderline range on the somatic problems subscale, while 1 (5%) child in the TD group scored in the clinical or borderline range on the subscales attention problems and aggression as well as on the 'wide-band' factor externalizing behaviours.

In relation to the Child Domain of the PSI, the findings revealed significant differences in the subscales of adaptability [ $F(2, 58) = 20.18, P < 0.001, \eta^2 = 0.53$ ], acceptability [ $F(2, 58) = 19.62, P < 0.001, \eta^2 = 0.5$ ], demandingness [ $F(2, 58) = 5.04,$

**Table 2.** Mean scores\* and standard deviations (SD) on the CBCL

$P = 0.012, \eta^2 = 0.2$ ], mood [ $F(2, 58) = 13.76, P < 0.001, \eta^2 = 0.43$ ] and distractability/hyperactivity [ $F(2, 58) = 6.46, P = 0.004, \eta^2 = 0.26$ ] as well as in the total Child Domain [ $F(2, 58) = 22.44, P < 0.001, \eta^2 = 0.56$ ]. Specifically, children with WS received significantly higher rates compared with both children with DS and TD children in the subscales of adaptability, acceptability, mood and the total Child Domain. Moreover, children with WS received significantly higher rates than TD children in the subscales of demandingness and distractability/hyperactivity. Children with DS received significantly higher rates compared with TD children in the subscale of acceptability and the total Child Domain. Mean scores and standard deviations on the subscales of the Child Domain are presented in Table 3.

The ANOVA and the *post hoc* test did not reveal any group differences in any of the subscales of the Parent Domain or the Life Stress Index of the PSI. Nevertheless, mothers of children with WS reported significantly higher scores in the Total Stress Index compared with mothers in the control group [ $F(2, 58) = 5.13, P = 0.011, \eta^2 = 0.47$ ]. Mean scores and standard deviations on the subscales of the Parent Domain and the Life Stress Index as well as the Total Stress Index of the PSI are presented in Table 4.

The percentage of mothers reporting scores into the clinical or borderline range on the Total Stress Index, the Child Domain and the Parent Domain of the PSI was also determined by comparing the raw scores with the standard scores. According to the results, in the WS group 12 (60%) mothers scored in the clinical or borderline range on the Total Stress Index, 8 (40%) on the Child Domain and 8 (40%) on the Parent Domain. On the other hand, only 3 (15%) mothers in the DS group and 2 (10%) mothers in the TD group scored in the clinical or borderline range on the Total Stress Index. Moreover, only 5 (25%) mothers in the DS group and 5 (25%) mothers in the TD group

**Table 3.** Mean scores\* and standard deviations (SD) on the Child Domain of the Parenting Stress Index

	DS		WS		TD	
	Mean	SD	Mean	SD	Mean	SD
Adaptability	21.93	2.84	28.85	5.13	18.33	4.50
Acceptability	13.50	3.41	17.77	4.36	8.92	2.50
Demandingness	17.36	3.41	18.54	5.59	13.33	3.47
Mood	8.93	1.98	12.85	2.61	8.50	2.36
Distractability/hyperactivity	23.21	5.74	27.23	3.98	20.00	5.20
Reinforcement for parents	11.36	2.37	11.69	2.36	10.50	2.11
Total Child Domain	96.29	10.77	116.92	16.19	79.58	14.7

\*Raw scores.

DS, Down syndrome; TD, typically developing; WS, Williams syndrome.

**Table 4.** Mean scores\* and standard deviations (SD) on the Parent Domain of the Parenting Stress Index, the Life Stress and the Total Stress Indices

	DS		WS		TD	
	Mean	SD	Mean	SD	Mean	SD
Depression	22.64	3.93	21.92	5.81	21.17	7.08
Attachment	17.29	14.62	13.46	2.60	13.00	3.33
Role restriction	20.14	2.83	21.00	4.93	20.33	6.59
Sense of competence	39.71	6.69	38.77	6.15	37.83	6.85
Social isolation	11.50	2.59	12.62	4.33	11.50	5.55
Spouse relationship	14.93	3.93	18.08	3.77	16.75	5.63
Parental health	16.21	3.07	16.08	3.25	15.17	2.55
Total Parent Domain	142.43	23.43	141.92	18.08	135.75	28.56
Life Stress Index	7.43	5.46	10.08	8.49	4.75	6.84
Total Stress Index	238.71	31.36	258.85	29.30	215.33	40.91

\*Raw scores.

DS, Down syndrome; TD, typically developing; WS, Williams syndrome.

scored in the clinical or borderline range on the Parent Domain, while none of these mothers scored in this range on the Child Domain.

In addition, the 'wide-band' factors and the total CBCL score were correlated with the Child Domain, the Parent Domain, the Life Stress Index and the Total Stress Index of the PSI. The findings showed that in the WS and the DS group, the externalizing behaviours were significantly positively correlated with the total Child Domain ( $r = 0.689, P = 0.013$  and  $r = 0.631, P = 0.015$  respectively). However, the results showed a significant positive correlation between externalizing behaviours and Total Stress Index ( $r = 0.535, P = 0.003$ ) and a significant negative correlation between internalizing behaviours and Life Stress ( $r = -0.561, P = 0.001$ ) in the WS group, but not in the DS group. On the other hand, in the control group internalizing behaviours, externalizing behaviours as well as the total CBCL score were significantly positively correlated with the Child Domain ( $r = 0.811, P = 0.001, r = 0.867, P < 0.001$  and  $r = 0.875, P < 0.001$  respectively), the Parent Domain ( $r = 0.718, P = 0.009, r = 0.662, P = 0.019$  and  $r = 0.737, P = 0.006$  respectively), the Life Stress

Index ( $r = 0.852, P < 0.001, r = 0.839, P = 0.001$  and  $r = 0.841, P = 0.001$  respectively) and the Total Stress Index ( $r = 0.793, P = 0.002, r = 0.773, P = 0.003$  and  $r = 0.829, P = 0.001$  respectively).

## Discussion

The present study investigated the behavioural profile and its relation to maternal stress in Greek young children with WS in comparison with young children with DS and matched controls. According to their mothers' ratings, young children with WS exhibit more emotional problems, fears and anxieties than young children with DS and TD children. In particular, it was reported that children with WS show difficulty in modulating emotions and they often fluctuate from happiness to sadness, while sometimes they may seem unhappy or grieved without an apparent cause. Another referred problem is the fear of failure and low frustration tolerance. Mothers also characterized their children with WS as 'tense', 'anxious' and 'worried'. Children with WS can become extremely upset and respond excessively if their wishes are denied or if they do not receive constant reassurance for their performance. Moreover, children with WS are often restless and they could be devastated by minor or unexpected changes in routine or schedule, which seem commonplace to others. It was also reported that children with WS are strongly attached to adults and express fear in new situations. Emotional instability, anxiety and depression are classified as 'internalizing' maladaptive behaviours (Achenbach & McConaughy 1992; Dykens *et al.* 2000). Distractability and attentional problems, which are considered as core deficiencies in WS, were also found in the present study. Mothers reported that visual or auditory background stimulation may cause WS children to lose concentration, which in turn results in failure to complete tasks.

One finding that deserves particular attention is that in this study children with WS are described as more withdrawn compared with TD children. At first glance this result contrasts with

results reported in previous studies, which demonstrate that children with WS are more gregarious and empathic than TD children (Gosch & Pankau 1994; Klein-Tasman & Mervis 2003; Doyle *et al.* 2004). However, this finding could be explained considering the items that comprise the subscale Withdrawn in the CBCL as well as the scores obtained on each item. The subscale Withdrawn in the CBCL consists of the following items: (i) acts too young for age (item 2); (ii) avoids looking others in the eye (item 4); (iii) does not answer when people talk to him/her (item 23); (iv) refuses to play active games (item 62); (v) seems unresponsive to affection (item 67); (vi) shows little affection towards people (item 70); (vii) shows little interest in things around him/her; and (viii) withdrawn, does not get involved with others. The results showed that children with WS received higher ratings compared with TD children in the items 2, 4, 23, 62, 71. Indeed there are several indications that children with WS may show deficiencies in affective attunement and reciprocal social interactions (Klein-Tasman *et al.* 2007). Given that individuals with WS have developmental delays (Mervis & Klein-Tasman 2000), they generally show remarkable deficits in communicative and non-communicative abilities (Mervis & Bertrand 1997; Mervis *et al.* 2000). It has been observed that although children with WS show intense interest in others' faces, even from a very early age, their eye contact patterns are qualitatively different and less well modulated than the eye contact patterns of TD children, and are characterized as abnormal (Udwin & Yule 1991; Mervis & Klein-Tasman 2000). Klein-Tasman and colleagues (2007) found that approximately 30% of their sample showed abnormality in the behaviour Response to Name. Moreover, it is reported that children with WS are usually rejected by their peers and are socially isolated (Einfeld *et al.* 2001). The finding that children with WS refuse to play active games more often than TD children may be explained by their deficiencies in fine and gross motor skills characterizing the condition. Individuals in this population exhibit low muscle tone as well as difficulties in visuo-motor co-ordination and orientation, which may prevent them from participating in such activities. However, in accordance with previous studies, children with WS in this study did not receive high ratings in the items 67, 78 and 98, which are related to the expression of affection and social involvement.

The behavioural profile of Greek young children with WS that emerged in the present study is in agreement with findings from other studies that examined young children, adolescents and adults with WS coming from western cultures such as German, the USA and the Netherlands as well as Asian cultures such as Japan (Gosch & Pankau 1997; Sarimski 1997; Van Lie-

shout *et al.* 1998; Fidler *et al.* 2000). Thus, the consistency of the socio-emotional characteristics of children with WS across cultures and developmental stages indicates a strong influence of the genetic phenotype.

Nevertheless, it is noteworthy that in this study only 25% of children with WS did fall into the clinical or borderline range in the total CBCL score. In contrast, in the studies of Sarimski (1997) and Fidler and colleagues (2000) 71% and 75% of children with WS respectively fell in the clinical or borderline range in the total CBCL score. This discrepancy may be explained by the Greek mothers' confined acquaintance with the behavioural characteristics of WS or by the age differences of the participants across studies. In Greece parents are informed about the diagnosis of WS only by the geneticist, who usually emphasizes the medical problems of the condition. On the other hand, there are no psychological services or organizations that specialize on the syndrome. As mentioned before, only 4 (20%) of the children with WS follow intervention programs for modification of maladaptive behaviours. Thus, mothers have usually very limited knowledge about the behavioural and socio-emotional characteristics of the condition. As a result mothers may attribute behaviours typical of WS to the child's intention, and not to the genetic abnormality. In other words, mothers may fail to acknowledge the severity and impact of WS behaviours, because they consider them as personality traits. Similar attitudes towards their child's behaviour are reported by parents of children with ADHD, a developmental disorder caused by biological abnormalities (Maniadaki *et al.* 2007). Another explanation lies in the way mothers scored the questionnaires, which may reflect their tendency to avoid the stigma of having a 'different' child.

The difference between the present and previous studies in the percentage of children who are reported to fall into the clinical or borderline range may also be accounted for by differences in the age range of the samples. In Sarimski's (1997) and Fidler and colleagues (2000) studies participants were toddlers as well as school children. Specifically, in Sarimski's study the age range of the participants was 1 to 12 years (mean age 6.7 years) and in Fidler and colleagues (2000) study it was 3–10 years (mean age 6.1 years). On the other hand, the present study focuses on young children and the age range of the participants was 3–6 years (mean age 5.1 years). Although the distribution of the participants at different ages is not reported in the previous studies, it is possible that mothers of older children score higher in the items regarding socio-emotional problems, because these problems may be brought out more intensely as the child enters school, and affect his academic and social functioning. On the contrary, mothers of

young children may feel that their child's socio-emotional problems will improve as the child grows older or can be changed through appropriate upbringing practices.

Another important finding is that mothers of children with WS, but not mothers of children with DS, report higher levels of stress compared with mothers of TD children, which was mainly related with adaptability and mood problems. Similarly, Sarimski (1997) found that mothers in the WS group reported higher levels of stress compared with the normal distribution. In addition, the present study demonstrated that the number of mothers in the WS group who reported stress levels in the clinical or borderline range was fourfold compared with the DS group and sixfold compared with the TD group. Correlational statistics showed that maternal stress seems to increase as the child exhibits more externalizing behaviours, but it seems to decrease as the child exhibits more internalizing behaviours. In accordance to our finding, other studies demonstrated that maternal stress and family problems in WS groups are associated with externalizing behaviours (Van Lieshout *et al.* 1998; Fidler *et al.* 2000). This finding acquires more interest when one considers the fact that internalizing behaviours consist a salient problem in this condition. However, it may be accounted for by maternal attitudes towards internalizing and externalizing behaviours. One could hypothesize that mothers perceive externalizing behaviours as more threatening for the normal functioning of the home, than internalizing behaviours, and feel less capable of dealing with them. In other words, externalizing behaviours may diminish mothers' sense of self-efficacy causing in turn elevated levels of stress. Moreover, externalizing behaviours are highly opposed to Greek cultural norms that emphasize obedience and respect (Maniadaki *et al.* 2005, 2007). This finding may be important for clinical practice. The success of any intervention program for modification of maladaptive behaviours presupposes mothers' cooperation, as they mediate for the application of its basic principles at home. Thus, it is important for the specialist to know not only the child's behavioural profile, but also mothers' perceptions about these behaviours.

The present study examined the association between maternal stress and maladaptive behaviours in WS using standardized questionnaires. Nevertheless, this issue deserves further investigation through direct observations of mothers' spontaneous reactions towards children's genetically originated behaviours. Moreover, maternal reactions should be related to certain aspects of culture as well as their upbringing practices. In this way it would be possible to gain a more thorough understanding of the nature vs. nurture interaction.

## Key messages

### Practice

- It seems that emotional problems as well as anxiety and depression are syndrome-specific characteristics in WS.
- Mothers of children with WS seem to experience more stress compared with mothers of TD children.
- Greek mothers of children with WS tend not to assess their children into the clinical range.

### Policy

- Intervention strategies need to take into account mothers' perceptions about their child's maladaptive behaviours.

### Research

- In future studies it would be beneficial to investigate maternal spontaneous behaviours during communicative exchanges with their child.

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